General

Guideline Title

Post-polio syndrome.

Bibliographic Source(s)

Farbu E, Gilhus NE, Barnes MP, Borg K, de Visser M, Howard R, Nollet F, Opara J, Stalberg E. Post-polio syndrome. In: Gilhus NE, Barnes MP, Brainin M, editor(s). European handbook of neurological management. 2nd ed. Vol. 1. Oxford (UK): Wiley-Blackwell; 2011. p. 311-9. [99 references]

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Farbu E, Gilhus NE, Barnes MP, Borg K, de Visser M, Driessen A, Howard R, Nollet F, Opara J, Stalberg E. EFNS guideline on diagnosis and management of post-polio syndrome. Report of an EFNS task force. Eur J Neurol 2006 Aug;13(8):795-801.

Recommendations

Major Recommendations

The levels of evidence (Class I-IV) supporting the recommendations and ratings of recommendations (A-C, Good Practice Points) are defined at the end of the "Major Recommendations" field.

Diagnostic Criteria

As post-polio syndrome (PPS) is reckoned to be a chronic disease, the European Federation of Neurological Societies (EFNS) task force on post-polio syndrome recommends that the criteria published by March of Dimes (MoD) in 2000 should be regarded as universal criteria for PPS (refer to the original guideline document for details). The diagnosis of PPS is an exclusion diagnosis with no test or analysis specific for PPS, and the role of the investigation is to rule out every other possible cause for the new symptoms and clinical deterioration.

Therapeutic Interventions

Level A Recommendations

- Some controlled studies of potential specific medical treatments for PPS have been completed, and no definitive therapeutic effect has been reported for the agents pyridostigmine, steroids, amantadine, modafinil, and coenzyme Q10.

Level B Recommendations

- Supervised muscular training, both isokinetic and isometric, is a safe and effective way to prevent further decline of muscle strength in slightly or moderately weak muscle groups and can even reduce symptoms of muscular fatigue, muscle weakness and pain in selected post-polio patients. A prolonged effect up to one year after well-defined training programmes has been reported.
- There are no studies evaluating the effect of muscular training in patients with severe weakness and the long-term effect of such training is not yet explored.
- Precautions to avoid muscular overuse should be taken with intermittent breaks, periods of rest between series of exercises and submaximal work load.
- Training in a warm climate and non-swimming water exercises are particularly useful.

Level C Recommendations

- Recognition of respiratory impairment and early introduction of non-invasive ventilatory aids prevent or delay further respiratory decline and the need of invasive respiratory aids.
- Respiratory muscle training can improve pulmonary function.
- Group training, regular follow-ups, and patient education are useful for the patients' mental status and well-being.
- Lightweight carbon orthoses can be more proper than metal orthoses.

Good Practice Points

- Weight loss and adjustment and introduction of properly fitted assistive devices is helpful, but lack significant scientific evidence.

Definitions:

Evidence Classification Scheme for a Diagnostic Measure

Class I: A prospective study in a broad spectrum of persons with the suspected condition, using a 'gold standard' for case definition, where the test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class II: A prospective study of a narrow spectrum of persons with the suspected condition, or a well-designed retrospective study of a broad spectrum of persons with an established condition (by 'gold standard') compared to a broad spectrum of controls, where test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class III: Evidence provided by a retrospective study where either persons with the established condition or controls are of a narrow spectrum, and where test is applied in a blinded evaluation

Class IV: Any design where test is not applied in blinded evaluation OR evidence provided by expert opinion alone or in descriptive case series (without controls)

Evidence Classification Scheme for a Therapeutic Intervention

Class I: An adequately powered prospective, randomized, controlled clinical trial with masked outcome assessment in a representative population or an adequately powered systematic review of prospective randomized controlled clinical trials with masked outcome assessment in representative populations. The following are required:

a. Randomization concealment
b. Primary outcome(s) is/are clearly defined
c. Exclusion/inclusion criteria are clearly defined
d. Adequate accounting for dropouts and crossovers with numbers sufficiently low to have minimal potential for bias
e. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences

Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a–e above or a randomized, controlled trial in a representative population that lacks one criteria a–e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (established as probably useful/predictive or not useful/predictive) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (established as possibly useful/predictive or not useful/predictive) requires at least two convincing class III studies.

Rating of Recommendations for a Therapeutic Intervention

Level A rating (established as effective, ineffective, or harmful) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (probably effective, ineffective, or harmful) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (possibly effective, ineffective, or harmful) requires at least two convincing class III studies.

Good Practice Point When only class IV evidence was available but consensus could be reached the task force gives recommendations as Good Practice Points.

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Post-polio syndrome (PPS)

Guideline Category

Diagnosis

Management

Rehabilitation

Treatment

Clinical Specialty

Family Practice

Internal Medicine

Neurology

Physical Medicine and Rehabilitation

Intended Users

Physical Therapists

Physicians

Respiratory Care Practitioners

Guideline Objective(s)

To revise the existing European Federation of Neurological Societies (EFNS) task force document, with regard to a common definition of post-polio syndrome (PPS) and evaluate the existing evidence for the clinical effectiveness of therapeutic interventions and on this basis provide updated clinical guidelines for management of PPS

Target Population

Patients with post-polio syndrome (PPS)

Interventions and Practices Considered

Diagnosis

Assessing symptoms and ruling out all other possible causes of new symptoms

Treatment/Management

1. Supervised muscular training (isokinetic and isometric)
2. Training in a warm climate, non-swimming water exercises
3. Precautions to avoid muscle overuse
4. Use of non-invasive ventilatory aids
5. Respiratory muscle training
6. Group training, regular follow-up, and patient education
7. Weight loss
8. Use of properly fitted assistive devices

Note: Pyridostigmine, steroids, amantadine, modafinil, and coenzyme Q were considered but not recommended because of lack of therapeutic effect.

Major Outcomes Considered

Effectiveness of treatment in improving muscle strength and cardiovascular fitness and reducing pain

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Medline via PubMed, EMBASE, ISI and the Cochrane Library were searched with time limits 1966 until 2004. Search terms were post-polio syndrome, post-poliomyelitis, PPMA, PPMD, poliomyelitis in combination with management, therapy, treatment, medicaments, physiotherapy and intervention.

In the present revised document, the database search was supplied with the years 2004–2009.

No meta-analyses of interventions for post-polio syndrome (PPS) were found when searching the databases, but one Cochrane review is being

prepared.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Evidence Classification Scheme for a Diagnostic Measure

Class I: A prospective study in a broad spectrum of persons with the suspected condition, using a 'gold standard' for case definition, where the test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class II: A prospective study of a narrow spectrum of persons with the suspected condition, or a well-designed retrospective study of a broad spectrum of persons with an established condition (by 'gold standard') compared to a broad spectrum of controls, where test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class III: Evidence provided by a retrospective study where either persons with the established condition or controls are of a narrow spectrum, and where test is applied in a blinded evaluation

Class IV: Any design where test is not applied in blinded evaluation OR evidence provided by expert opinion alone or in descriptive case series (without controls)

Evidence Classification Scheme for a Therapeutic Intervention

Class I: An adequately powered prospective, randomized, controlled clinical trial with masked outcome assessment in a representative population or an adequately powered systematic review of prospective randomized controlled clinical trials with masked outcome assessment in representative populations. The following are required:

a. Randomization concealment
b. Primary outcome(s) is/are clearly defined
c. Exclusion/inclusion criteria are clearly defined
d. Adequate accounting for dropouts and crossovers with numbers sufficiently low to have minimal potential for bias
e. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences

Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a–e above or a randomized, controlled trial in a representative population that lacks one criteria a–e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

Methods Used to Analyze the Evidence

Systematic Review

Description of the Methods Used to Analyze the Evidence

Data were classified according to their scientific level of evidence as Class I–IV (see the "Rating Scheme for the Strength of the Evidence" field).

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

Recommendations are given as Level A–C according to the scheme for European Federation of Neurological Societies (EFNS) guidelines (see the "Rating Scheme for the Strength of the Recommendations" field). When only Class IV evidence was available but consensus could be reached the Task Force gives recommendations as Good Practice Points (GPP). Consensus was reached mainly through e-mail correspondence.

A questionnaire about diagnosis, management and care of post-polio patients was answered by the group members from the Netherlands, Norway, Poland, Sweden, and United Kingdom in the first version; this has not been repeated in this revision.

Rating Scheme for the Strength of the Recommendations

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (established as probably useful/predictive or not useful/predictive) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (established as possibly useful/predictive or not useful/predictive) requires at least two convincing class III studies.

Rating of Recommendations for a Therapeutic Intervention

Level A rating (established as effective, ineffective, or harmful) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (probably effective, ineffective, or harmful) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (possibly effective, ineffective, or harmful) requires at least two convincing class III studies.

Good Practice Point When only class IV evidence was available but consensus could be reached the task force gives recommendations as Good Practice Points.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Peer Review

Description of Method of Guideline Validation

The guidelines were validated according to the European Federation of Neurological Societies (EFNS) criteria (see the "Availability of Companion Documents" field).

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for selected recommendations (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate diagnosis and treatment of post-polio syndrome (PPS)

Potential Harms

Precautions to avoid muscular overuse while exercising should be taken with intermittent breaks, periods of rest between series of exercises and submaximal work load.

Qualifying Statements

Qualifying Statements

This guideline provides the view of an expert task force appointed by the Scientific Committee of the European Federation of Neurological Societies (EFNS). It represents a peer-reviewed statement of minimum desirable standards for the guidance of practice based on the best available evidence. It is not intended to have legally binding implications in individual cases.

Implementation of the Guideline

Description of Implementation Strategy

The European Federation of Neurological Societies has a mailing list and all guideline papers go to national societies, national ministries of health, World Health Organisation, European Union, and a number of other destinations. Corporate support is recruited to buy large numbers of reprints of the guideline papers and permission is given to sponsoring companies to distribute the guideline papers from their commercial channels, provided there is no advertising attached.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Farbu E, Gilhus NE, Barnes MP, Borg K, de Visser M, Howard R, Nollet F, Opara J, Stalberg E. Post-polio syndrome. In: Gilhus NE, Barnes MP, Brainin M, editor(s). European handbook of neurological management. 2nd ed. Vol. 1. Oxford (UK): Wiley-Blackwell; 2011. p. 311-9. [99 references]

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2006 Aug (revised 2011)

Guideline Developer(s)

European Academy of Neurology - Medical Specialty Society

Source(s) of Funding

European Federation of Neurological Societies

Guideline Committee

European Federation of Neurological Societies Task Force on Post-Polio Syndrome

Composition of Group That Authored the Guideline

Task Force Members: E. Farbu, Stavanger University Hospital, Norway; N. E. Gilhus, University of Bergen, and Haukeland University Hospital, Bergen, Norway; M. P. Barnes, Hunters Moor Hospital, Newcastle upon Tyne, UK; K. Borg, Karolinska Intitutet/Karolinska Hospital, Stochkholm, Sweden; M. de Visser, University of Amsterdam, Amsterdam, The Netherlands; R. Howard, St Thomas' Hospital, London, UK; F. Nollet, University of Amsterdam, Amsterdam, The Netherlands; J. Opara, Repty Rehab Centre ul. Sniadeckio, Tarnowskie Góry, Poland; E. Stalberg, University Hospital, Uppsala, Sweden

Financial Disclosures/Conflicts of Interest

The authors have reported no conflicts of interests.

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Farbu E, Gilhus NE, Barnes MP, Borg K, de Visser M, Driessen A, Howard R, Nollet F, Opara J, Stalberg E. EFNS guideline on diagnosis and management of post-polio syndrome. Report of an EFNS task force. Eur J Neurol 2006 Aug;13(8):795-801.

Guideline Availability

Electronic copies: Available in Portable Document Format (PDF) from the European Federation of Neurological Societies (EFNS) Web site .

Availability of Companion Documents

The following is available:

- Brainin M, Barnes M, Baron JC, Gilhus NE, Hughes R, Selmaj K, Waldemar G; Guideline Standards Subcommittee of the EFNS Scientific Committee. Guidance for the preparation of neurological management guidelines by EFNS scientific task forces – revised recommendations 2004. Eur J Neurol. 2004 Sep;11(9):577-81. Electronic copies: Available in Portable Document Format (PDF) from the European Federation of Neurological Societies Web site .

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI on April 9, 2007. The information was verified by the guideline developer on May 15, 2007. This NGC summary was updated by ECRI Institute on February 20, 2012.

Copyright Statement

Disclaimer

NGC Disclaimer